**Scholars Journal of Medical Case Reports**
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

**Neurointensive Care**

# Subarachnoid Hemorrhage Revealing a Moyamoya Disease

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**DOI:** https://doi.org/10.36347/sjmcr.2025.v13i06.032 | **Received:** 08.05.2025 | **Accepted:** 14.06.2025 | **Published:** 17.06.2025

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**Abstract** | **Case Report**

Moyamoya Disease (MMD) is a rare and extremely severe chronic cerebrovascular disorder characterized by progressive stenosis of the internal carotid artery and the development of an abnormal vascular network at the base of the brain. It presents clinically with ischemic strokes or hemorrhages. The case reported is that of a 47-year-old woman admitted for a subarachnoid hemorrhage. Imaging revealed a bilateral form of MMD. The patient received supportive medical treatment (mechanical ventilation, nimodipine) and an external ventricular drainage procedure, but the outcome was unfavorable due to the occurrence of cerebral ischemia, ultimately resulting in death. This case illustrates a rare presentation of MMD and highlights the severity of the disease progression in the absence of early revascularization, as well as the importance of prompt diagnosis and specialized intervention.

**Keywords:** Moyamoya Disease (MMD), Subarachnoid hemorrhage, Cerebrovascular disorder, Revascularization surgery, Cerebral ischemia.



## INTRODUCTION

Moyamoya disease is a chronic cerebrovascular condition characterized by progressive stenosis of the terminal portion of the internal carotid artery and the development of an abnormal vascular network at the base of the brain. Clinical symptoms may be related to either ischemic or hemorrhagic stroke. The diagnosis is established through cerebral angiography. Early intervention is essential and primarily involves revascularization surgery. We report the case of a patient diagnosed with Moyamoya disease following a subarachnoid hemorrhage.

## MEDICAL OBSERVATION

This is a 47-year-old female patient with no known past medical history and no recent antibiotic use, who was admitted to the emergency department for a non-febrile deterioration of consciousness. The history of the illness dates back seven days prior to admission, with the sudden onset of intense headaches that were resistant to symptomatic treatment. No seizure activity was reported by the patient's family. On admission, her Glasgow Coma Scale (GCS) score was 10 (E2, V3, M5). There were no sensorimotor deficits, pupils were equal and reactive, neck stiffness was noted. Blood pressure was 125/76 mmHg, heart rate 85 bpm, respiratory rate 22 breaths/min, $SpO_2$ 97% on room air, capillary blood glucose 1.3 g/L, and body temperature was 37.3°C.

The patient was admitted to the intensive care unit, and intubation was performed based on neurological criteria. Given the sudden, non-traumatic, and afebrile onset of consciousness disturbance, an emergency brain CT scan was performed, revealing a Fisher grade 4 subarachnoid hemorrhage with intraventricular flooding but without hydrocephalus (Evans Index: 0.2). A subsequent cerebral CT angiography showed findings suggestive of a saccular aneurysm of the left thalamoperforating artery and a probable fusiform aneurysm of the M1 segment of the middle cerebral artery. Due to the presence of an aneurysm on CT angiography, a cerebral arteriography was performed. It revealed bilateral diffuse narrowing of both cervical and intracranial internal carotid arteries, extending from the post-bulbar segment to the origin of the ophthalmic arteries, with complete occlusion just downstream. An anastomotic collateral network characteristic of bilateral Moyamoya disease was observed. No vascular malformations or signs suggestive of vasospasm were seen (Image 1).

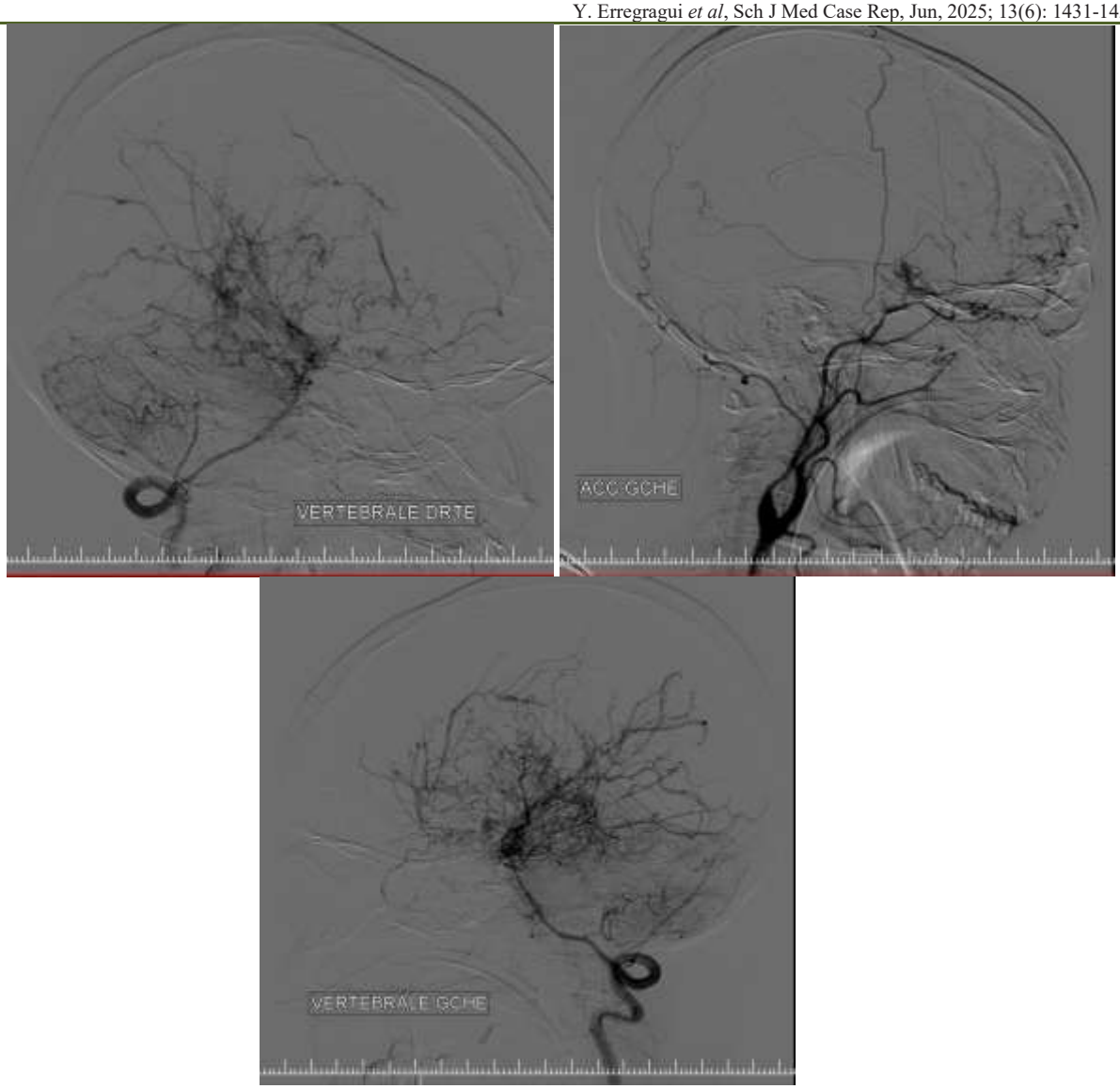


**Image 1: Cerebral arteriography showing stenosis of the internal carotid artery with the development of a Moyamoya-type collateral anastomotic network**

**Timeline of clinical evolution of the patient**

| Day | Event |
|---|---|
| 0 (admission) | 47-year-old woman with no medical history, admitted for non-traumatic, afebrile disturbance of consciousness.<br>**Clinical examination:** GCS score of 10, neck stiffness, no sensorimotor deficit, WFNS grade 4.<br>**CT angiography:** Suspicion of a left thalamoperforating artery aneurysm and middle cerebral artery involvement.<br>**Cerebral angiography:** Bilateral stenosis of the internal carotid arteries with a Moyamoya-type collateral network. |
| 4 | Placement of an external ventricular drain (EVD) for active hydrocephalus. |
| 9 | **Neurological deterioration with bilateral mydriasis.**<br>**Brain CT scan:** diffuse cerebral edema and extensive ischemic lesions. |
| 13 | Death of the patient |

No revascularization procedure was performed due to the acute context of subarachnoid hemorrhage and persistent impairment of consciousness. Therapeutic management focused on the prevention of secondary systemic brain insults, control of intracranial hypertension through deep sedation, appropriate head positioning (neutral, elevated at 30°), maintenance of low ventilatory pressures targeting a $PaCO_2$ of 35–38 mmHg, prevention of delayed cerebral ischemia using nimodipine, and strict blood pressure control with a systolic arterial pressure target between 120 and 140

mmHg. Neurological monitoring was carried out both clinically and with transcranial Doppler.

On day 4 of hospitalization, following a sedation weaning trial, the patient developed generalized tonic-clonic seizures, which resolved after administration of 2 mg of midazolam. Transcranial Doppler showed an increased pulsatility index (PI) of 1.42 and diastolic velocity (Vd) of 24 cm/s on the right, and a PI of 1.37 with Vd of 26 cm/s on the left, prompting the resumption of sedation and a follow-up brain CT scan. The imaging revealed active tetraventricular hydrocephalus, leading to the placement of an external ventricular drain (EVD). The pressure level was set at 15 mmHg, with a drainage target of approximately 150 mL over 24 hours.

The clinical course was further complicated by the onset of bilateral mydriasis. A repeat CT scan showed diffuse ischemic lesions and widespread cerebral edema, ultimately leading to the patient's death on day 13.

## DISCUSSION

Moyamoya disease (MMD) is a rare cerebrovascular condition characterized by progressive stenosis of the intracranial portion of the internal carotid artery (ICA) and its proximal branches. This occlusion leads to the development of a fine vascular network at the base of the brain. Although numerous studies have expanded our understanding of MMD's clinical features, etiology, and pathophysiology, further investigation remains necessary to improve long-term outcomes through a better comprehension of this disease and its therapeutic approaches.

MMD is more frequently reported in patients from Asian countries, particularly Japan, Korea, and China [1,2]. A slight male predominance is noted, with two age peaks in distribution: the first between 5 and 9 years, and the second between 45 and 49 years [3].

The pathogenesis of MMD remains unclear. Histopathological studies have shown that the diameter of the carotid artery is significantly reduced in affected patients. Additionally, fibrous intimal thickening, irregular undulation of the internal elastic lamina, and attenuation of the media have been observed [4]. These changes result in arterial lumen narrowing, which may lead to ischemic stroke. Some studies have demonstrated elevated levels of growth factors and cytokines in the cerebrospinal fluid of MMD patients, including fibroblasts [5], intercellular adhesion molecule-1, E-selectin, cellular retinoic acid-binding proteins, and hepatocyte growth factor [6,7]. These angiogenic proteins may contribute to the formation of collateral circulation following indirect bypass revascularization. A genetic predisposition has also been strongly suggested, with numerous mutations implicated in the development of Moyamoya disease.

Clinical features of Moyamoya disease differ between children and adults. Most children present with transient ischemic attacks (TIA) or cerebral infarction, whereas approximately half of adult patients develop intracranial hemorrhage, and the other half may experience TIA, infarction, or both [4]. MMD usually causes cerebral ischemia in the internal carotid artery territory, manifesting as focal neurological signs such as dysarthria, aphasia, hemiparesis, headache, or seizures.

However, atypical symptoms such as syncope, paraparesis, visual disturbances, involuntary movements, intellectual disability, and rarely cognitive dysfunctions (e.g., short-term memory loss, irritability, or agitation) may also occur [9,10]. In the pediatric population, ischemic events are often triggered by hyperventilation (e.g., during crying or playing) [11]. In adults, about 50% of MMD cases are complicated by intracranial hemorrhage, primarily due to either rupture of dilated and fragile Moyamoya vessels or rupture of saccular aneurysms within the circle of Willis [12].

Diagnosis of MMD can be confirmed via imaging studies. Computed tomography (CT) may show small hypodense areas suggesting stroke in cortical regions, basal ganglia, white matter, or periventricular areas. However, CT may appear normal, especially in TIA cases.

CT angiography (CTA) can reveal stenoses, particularly involving the middle and anterior cerebral arteries.

Magnetic resonance imaging (MRI) helps localize ischemic or hemorrhagic lesions within the brain parenchyma. T1-weighted sequences can detect Moyamoya vessels in the basal ganglia and thalamus. T2*-weighted sequences may reveal asymptomatic microhemorrhages in 15–44% of adult patients. Magnetic resonance angiography (MRA) is also useful for non-invasive diagnosis by identifying stenoses at the distal internal carotid arteries.

Cerebral angiography remains the gold standard for diagnosing Moyamoya disease. It reveals stenosis or occlusion of the terminal ICA segment (C1–C2) and proximal segments of the anterior and middle cerebral arteries bilaterally. Stenosis or occlusion of the proximal posterior cerebral artery is seen in about 25% of cases. Arterial phase imaging shows a bilateral collateral network surrounding the obstructive lesions. These collaterals form a characteristic "puff of smoke" appearance— the hallmark of Moyamoya vasculature (Figure 1).

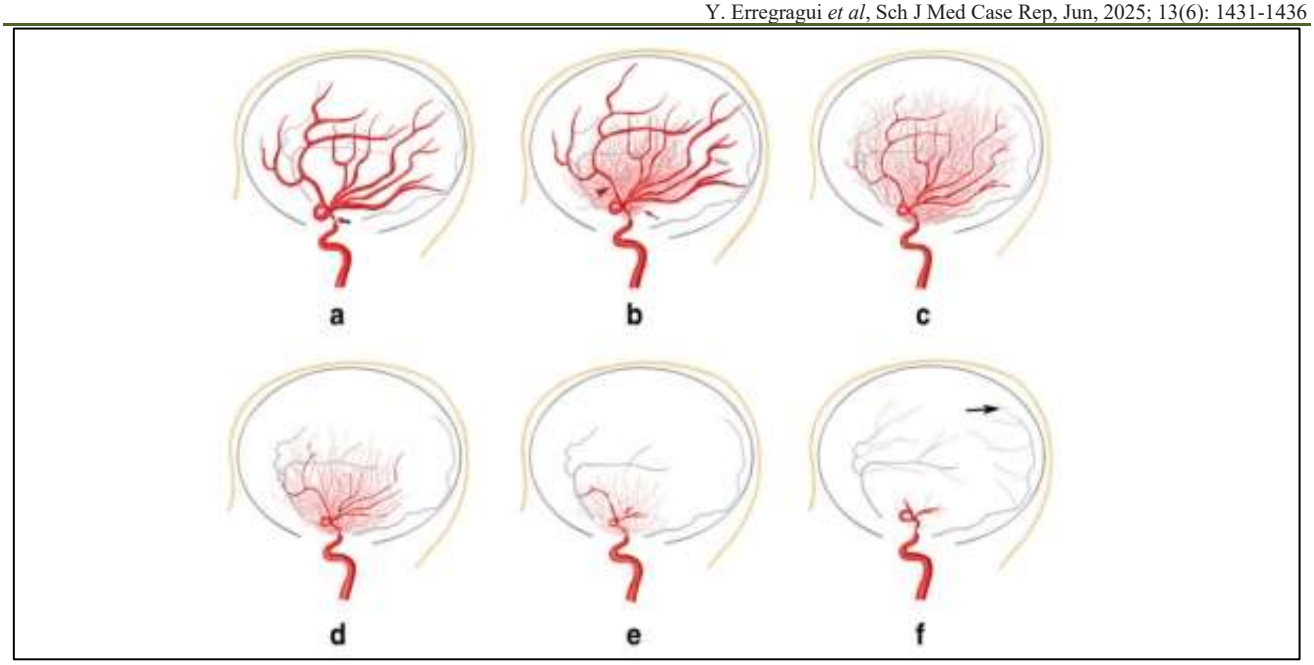


**Figure 1: Various arteriographic features of Moyamoya disease (13)**

The angiographic classification by Suzuki and Takaku is well known and widely used. It describes six grades of increasing disease severity (Table 1).

**Table 1: Angiographic classification by Suzuki and Takaku**

| Stade | Description |
|---|---|
| I | Sténose de la terminaison de l'artère carotide interne et de l'origine de A1 et M1. |
| II | Début de la formation du réseau de suppléance : sténose de l'ACI, dilatation des artères cérébrales antérieure, moyenne et postérieure en aval des lésions sténo-occlusives. |
| III | Intensification du réseau de suppléance. |
| IV | Apparition des anastomoses transdurales. |
| V | Réduction du réseau Moyamoya, occlusion des ACI, cérébrales antérieure, moyenne et postérieure. |
| VI | Disparition du réseau : suppléances cérébrales assurées par les anastomoses transdurales. |

Surgery remains, according to the current literature, the therapeutic option that has most consistently demonstrated efficacy in the management of patients with Moyamoya disease [8]. Its objective is to create or promote the development of collateral circulation in order to improve cerebral perfusion.

Two main categories of surgical cerebral revascularization techniques must be distinguished:

- **Direct techniques**, which provide immediate revascularization of the affected cerebral hemisphere, and
- **Indirect techniques**, which aim to induce neoangiogenesis by placing vascularized tissue in contact with the ischemic cerebral cortex.

Direct revascularization typically involves a termino-terminal anastomosis between a cortical branch of the middle cerebral artery (MCA) and the superficial temporal artery (STA), which is supplied by the external carotid artery (ECA)—an artery usually unaffected by stenosis in patients with Moyamoya disease.

In contrast, indirect methods consist of transposing various well-vascularized tissues—such as the STA, galea aponeurotica, dura mater, temporalis muscle, or omentum—into contact with the cortical surface. These procedures, collectively termed synangiosis, leverage the angiogenic capacity of these tissues to promote the gradual development of a functional collateral network. This neovascularization process generally occurs over a period of 1 to 6 months. Numerous technical variants have been described, depending on the tissues used.

Among them, EDAMS (encephalo-duro-arterio-myo-synangiosis) is considered the most comprehensive indirect technique. It involves the simultaneous transposition of a branch of the STA, the outer layer of the dura mater, and the temporalis muscle to maximize contact surface and angiogenic potential.

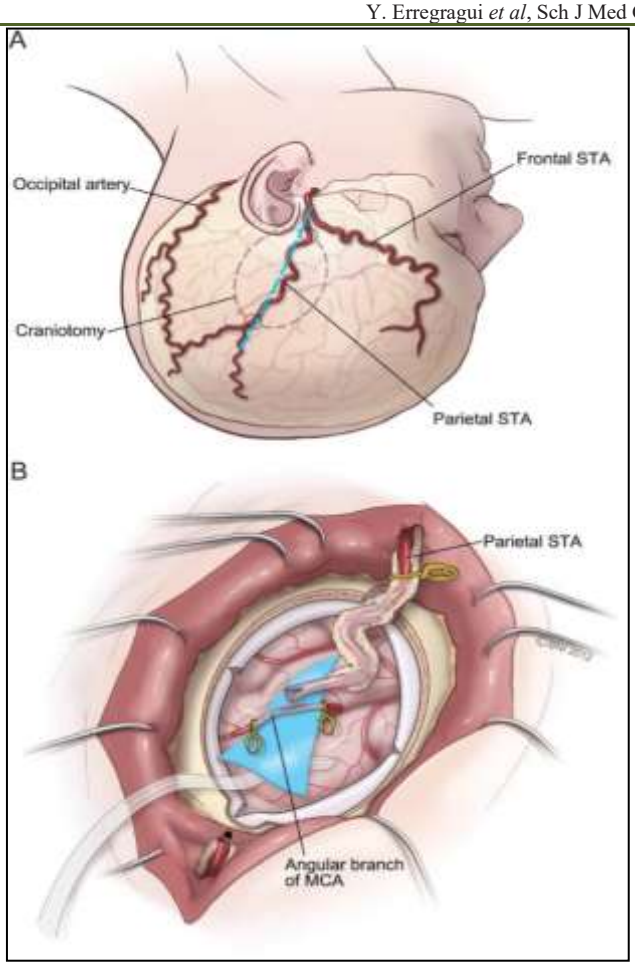


**Figure 2: Indirect surgical technique for the management of Moyamoya disease (MMD)**

The prognosis of Moyamoya disease remains guarded, particularly in cases of delayed diagnosis or the occurrence of complications, especially hemorrhagic events. Several factors have been associated with a poor outcome, including the occurrence of hemorrhagic stroke, female sex, familial forms of the disease, and symptom onset during childhood.

## CONCLUSION

Moyamoya disease is a rare vascular condition, often diagnosed at an advanced stage due to the variability and non-specificity of its clinical manifestations. Its discovery in the context of a subarachnoid hemorrhage—although less common than ischemic presentations—highlights the potential severity of this disease and the importance of early diagnosis. This case underscores the need to consider Moyamoya disease in the etiological workup of non-traumatic subarachnoid hemorrhage, particularly in young patients without obvious vascular risk factors. Appropriate neurosurgical management, combined with close follow-up, is essential to reduce the risk of hemorrhagic recurrence and to prevent disease progression.

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